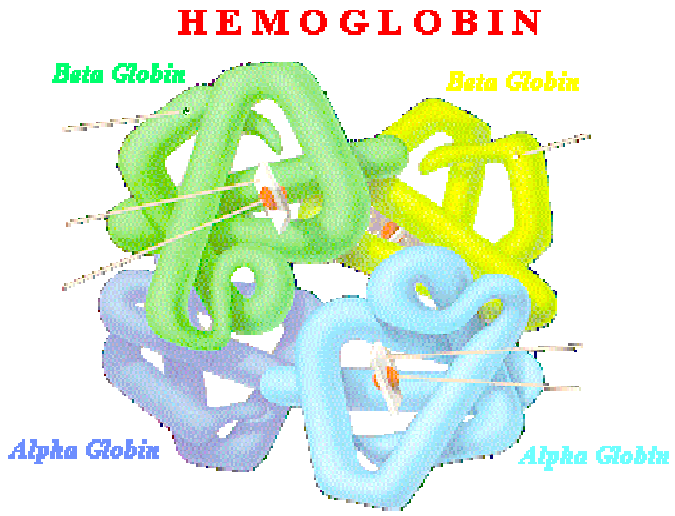
Hemoglobin Barts (F+ A+ Barts) Fact Sheet For Health Care Providers

In September 2001 the State of Utah began screening for hemoglobinopathies. Hemoglobinopathy screening identifies infants with sickle cell anemia, other non-sickle hemoglobin disorders, and hemoglobin traits.



Hemoglobin Bart's is a relatively common hemoglobin variant detected by High Performance Liquid Chromatography (HPLC) testing and is only seen during the newborn period. Infants with an alpha-thalassemia syndrome usually show hemoglobin Bart's on the newborn screening test.

Alpha-thalassemia is caused by deletions of the alpha globin genes on chromosome 16. Normal individuals have four copies of the alpha-globin gene, 2 on each chromosome. **The loss of one to four of these genes is possible**. The presence of hemoglobin Bart's on the newborn screen almost always indicates that one or more of the baby's alpha-globin genes are deleted. The severity of the alpha-thalassemia condition is directly related to the amount of gene deletions present. See table.

*Usual Genotypes	Alpha-Globin Gene Deletions	Clinical Features
αα/αα	0	Normal
-α/αα	1	Silent Carrier
--/αα or -α/-α	2	α-thalassemia trait
--/-α	3	Hb H Disease
--/--	4	Fetal Hydrops

* α indicates presence of α-globin gene. – indicates deletion of α-globin gene

Silent Carrier: ONE deleted alpha gene

This condition is clinically benign. If only one alpha gene is deleted, the other three genes can compensate nearly completely. These infants show normal CBC (Complete Blood Count) without microcytosis or anemia.

Alpha Thalassemia Trait: TWO deleted alpha genes

This condition, also referred to as alpha thalassemia minor, is characterized by microcytosis and hypochromia (low MCV, MCH) with *mild or no* anemia. The hypochromic anemia is often confused with iron deficiency anemia. Iron supplementation is ***not*** recommended unless the patient has a diagnosis of iron deficiency anemia.

Hemoglobin H Disease: THREE deleted alpha genes

Three deleted alpha genes generally result in a moderately severe form of alpha-thalassemia. Clinical manifestations of this disorder are variable, but most patients are anemic and develop some degree of splenomegaly. These patients should be followed by a pediatric hematologist.

Fetal Hydrops Syndrome: Four (all) alpha genes deleted

In fetal hydrops syndrome, also known as alpha thalassemia major, none of the alpha genes are present. This is not compatible with life and death usually occurs in utero or during early infancy.